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Ruptured Retroperitoneal Mucinous Cystadenocarcinoma with Synchronous Gastric Carcinoma and a Long Postoperative Survival: Case Report

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We describe an 86-year-old woman with a long survival following surgey for a massive retroperitoneal mucinous cystadenocarcinoma and a synchronous gastric carcinoma. Computed tomography showed a huge tumor with septation and calcification. Upper gastrointestinal radiography showed the additional gastric lesion. At operation, the 23 × 20 × 12-cm retroperitoneal tumor had ruptured. Tumor resection and distal gastrectomy including regional lymph nodes were performed. Mucinous peritoneal implants were removed as completely as possible. Histologically, the mucinous tumor showed limited invasion, whereas the poorly differentiated gastric adenocarcinoma showed no serosal invasion. Among 18 retroperitoneal mucinous cystadenocarcinomas reported in the English literature since 1965, only ours was associated with gastric carcinoma. Despite peritoneal implants, our patient has survived for 6 years without clinical recurrence. As at other sites, retroperitoneal mucinous cystadenocarcinoma often grows slowly. Total removal, even after peritoneal dissemination, can result in long survival.

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KEY WORDS: **mucinous cystadenocarcinoma; retroperitoneal; rupture; long survival; peritoneal implants**

INTRODUCTION

Retroperitoneal mucinous cystadenocarcinoma is very rare [1–13], and little is known concerning its pathogenesis, optimal treatment, and prognosis. We describe a case of retroperitoneal mucinous cystadenocarcinoma with long postoperative survival despite peritoneal implants and a synchronous gastric adenocarcinoma, in the context of relevant literature.

CASE REPORT

The patient was an 86-year-old woman with a complaint of abdominal tumor and no significant medical history. In February 1993, she was found to have an abdominal mass and was referred to our hospital. On physical examination, a massive, soft tumor was palpable, filling much of the abdomen. Laboratory data showed no abnormalities except for an elevated serum concentration of carcinoembryonic antigen (CEA), 66.1 ng/ml. The serum concentration of CA15-3 was normal. Ultrasonography (US) showed that the tumor was hypoechoic. On computed tomography (CT), the tumor had a low overall density and contained calcifications and septations (Fig. 1). No papillary nodules were demonstrated within the tumor. On angiography, no tumor blush or feeding artery was demonstrated. Additionally, a polypoid lesion on the lesser curvature was detected by upper gastrointestinal radiography performed due to high level of serum CEA (Fig. 2). An endoscopic biopsy specimen from the stomach showed poorly differentiated

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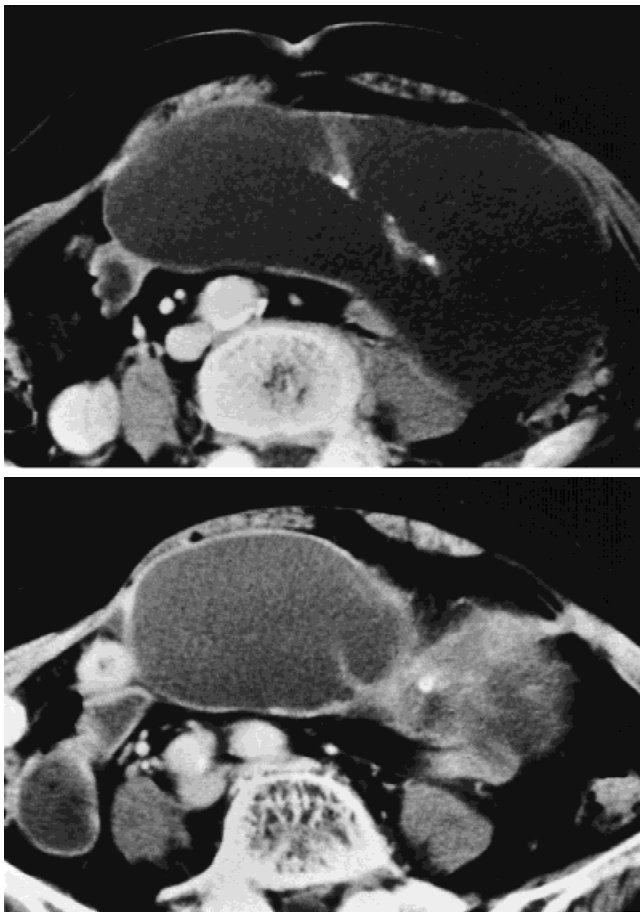

Fig. 1. CT. A large, generally low-density tumor shows calcification and septation.

adenocarcinoma. With a preoperative diagnosis of benign retroperitoneal cyst and carcinoma of the stomach, the patient underwent surgery in August 1993.

Laparotomy disclosed a massive tumor arising from the retroperitoneum. The tumor had ruptured, with mucinous peritoneal implants present nearby. The appendix, both ovaries, and the pancreas were grossly normal in size and shape. Resection of the retroperitoneal tumor and distal partial gastrectomy with regional lymph node dissection were performed. Peritoneal implants were removed as completely as possible.

The ruptured retroperitoneal tumor weighed 1,120 g and measured 23 × 20 × 12 cm. Macroscopically, the tumor was multilocular and filled with mucin. The internal surface appeared shaggy and showed fibrotic thickening, but no papillary nodules were visible (Fig. 3). Histologically, the cyst was lined by tall mucin-producing columnar epithelium showing a variety of atypical features, including a papillary growth pattern (Fig. 4). Minimal invasion of surrounding connective tissue was seen. In the mucinous peritoneal implants, similar-appearing tumor cells were seen floating in the mucin. A pathologic diagnosis of mucinous cystadenocarcinoma was made. No ectopic ovarian tissue was found. The pathologic diagnosis of the gastric lesion was poorly differentiated adenocarcinoma without serosal invasion. The histologic appearance of the retroperitoneal tumor differed completely from that of the gastric carcinoma. From these findings, we judged the cystic tumor to be a primary retroperitoneal mucinous cystadenocarcinoma. Histochemically neoplastic cells of both the carcinoma of the stomach and the retroperitoneal tumor showed sparse staining for CEA.

The postoperative course was uneventful. The serum concentration of CEA decreased to within normal limits. No adjuvant chemotherapy was administered. The patient is alive 6 years after operation with no signs of recurrence.

DISCUSSION

Including our patient, only 18 cases of retroperitoneal mucinous cystadenocarcinoma [1–13] have been reported in the English literature between 1965 and 1999 (Table I). With a single exception, patients have been female [10]. Ages of patients have ranged from 17 to 86 years. Reported symptoms of retroperitoneal mucinous cystadenocarcinoma typically were abdominal mass, distention, and discomfort or pain. As retroperitoneal tumors usually cause mild or nonspecific symptoms, discovery often is delayed. Maximum diameters of the tumors varied from 6 to 24 cm.

US and CT have facilitated detection of retroperitoneal tumors. While lymph node metastases or invasion of the surrounding connective tissue structures are evidence of malignancy, such findings have rarely been detected before operation. More specifically, while mucinous cystadenocarcinoma is difficult to differentiate preoperatively from benign mucinous cystadenoma [14], papillary nodules demonstrated within a cyst by US and CT have been reported to suggest malignancy [4].

The origin of retroperitoneal mucinous tumors is uncertain. While supernumerary ovary [1] has been considered a possible source of retroperitoneal mucinous cystic tumors, no ovarian tissue was found in our patient. A theory involving coelomic metaplasia [2,9,11] has gained increasing support as an explanation for ovarian epithelial neoplasms. A cystic invagination of the same coelomic epithelial layer with concurrent or subsequent metaplasia could account for retroperitoneal mucinous tumors. Mesothelial inclusion cysts have been reported in this location and residual mesothelium has been found in association with retroperitoneal mucinous cystadenocarcinoma [3]. These observations support the idea that retroperitoneal cystic tumors can develop through coelomic metaplasia involving a mesothelial cyst. This theory also permits occurrences in male patients.

As for treatment, total excision of the tumor has been performed in all reported cases (Table I). A total hyster-

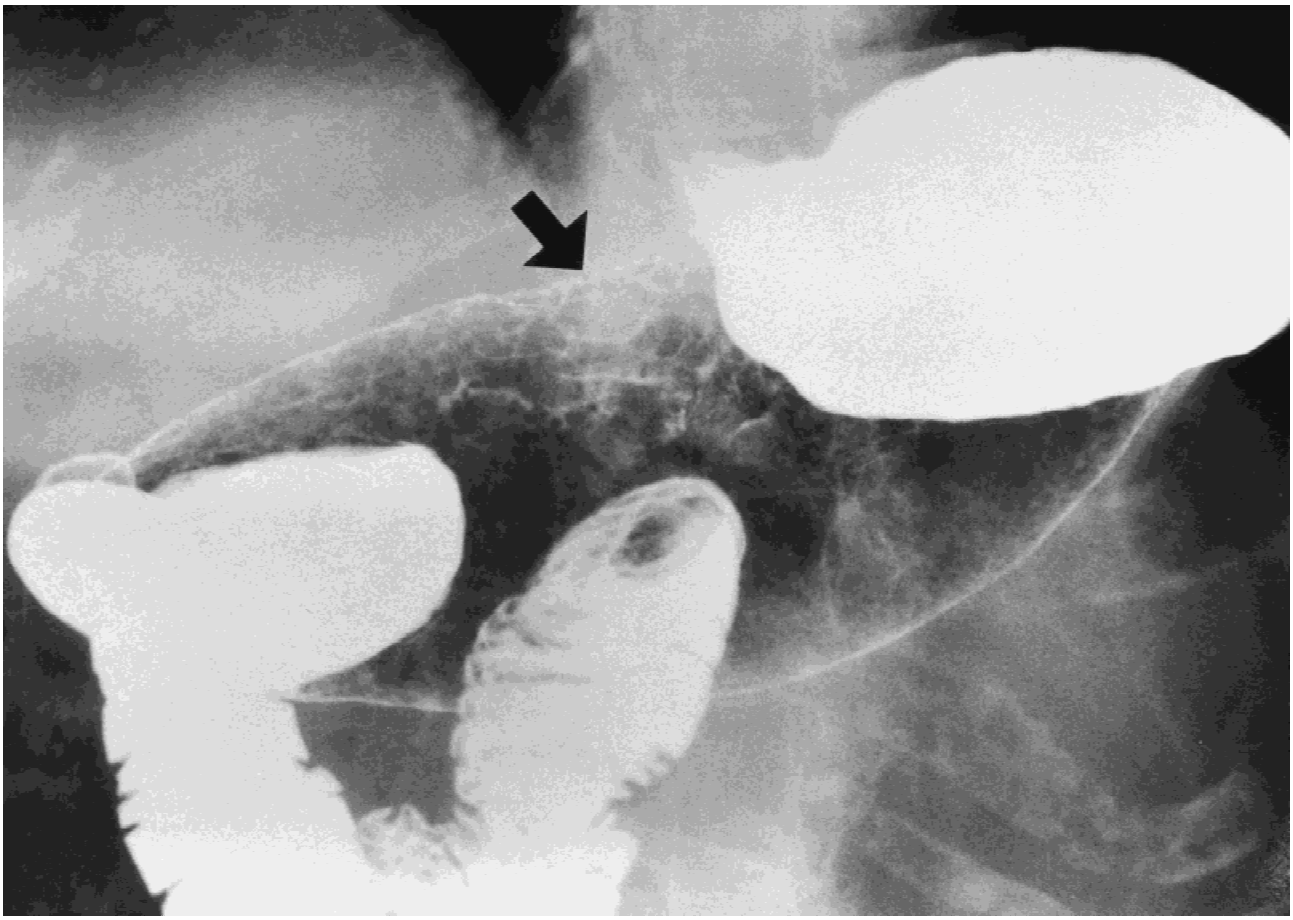

Fig. 2. Upper gastrointestinal radiograph shows polypoid lesion (arrow) of the lesser curvature.

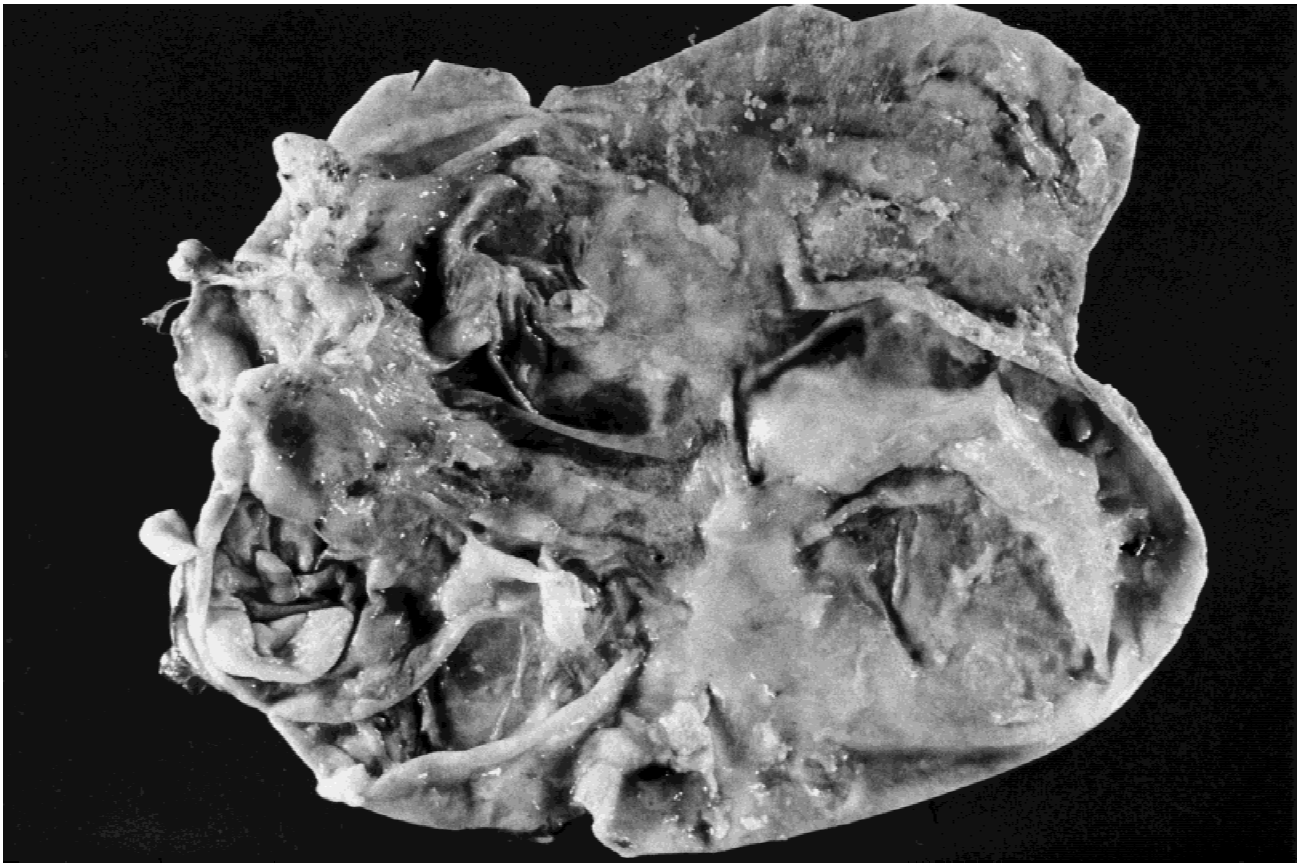

Fig. 3. Macroscopic appearance of the internal surface of the resected retroperitoneal tumor. The tumor is multilocular and filled with mucin, showing a shaggy inner surface and fibrosis.

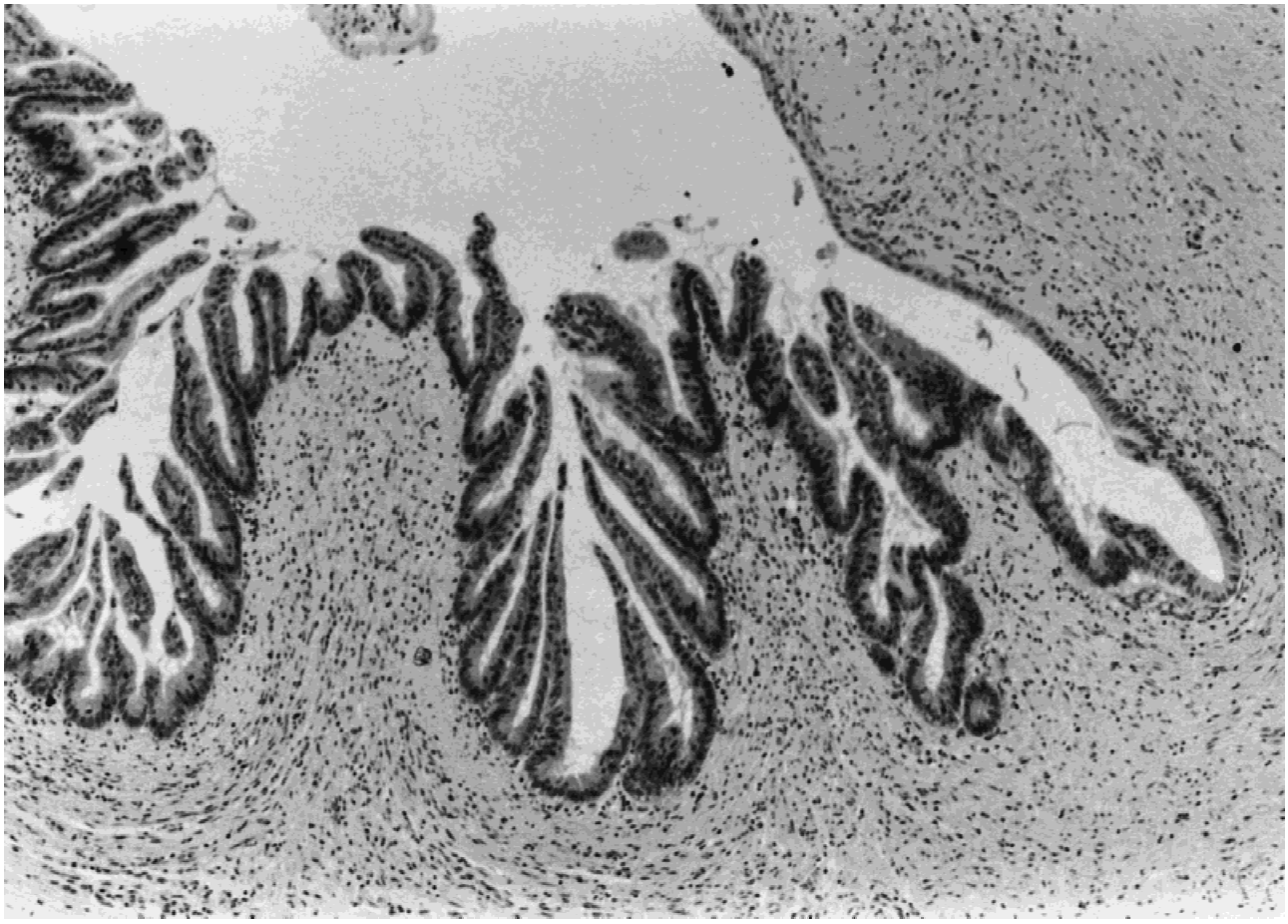

Fig. 4. Histologic findings in the retroperitoneal tumor, showing the cyst to be partly lined by papillary adenocarcinoma (hematoxylin-eosin, ×20).

TABLE I. Retroperitoneal Mucinous Cystadenocarcinomas Reported in English (1965–1999)*

Reference	Author	Sex	Age	Size	Surgical treatment	Combined therapy	Course
1	Roth and Ehrlich	F	48	550 g	Tumor excision	—	Spread and death 6 months after surgery
2	Storch and Raghavan	F	17	—	Tumor excision	Chemo	Paraovarian recurrence 21 months after surgery
3	Fujii et al.	F	69	23 cm	Tumor excision Hy, Adn	No	NER 36 months after surgery
4	Nelson et al.	F	35	20 cm	Tumor excision Hy, Adn	No	NER 22 months after surgery
5	Banerjee and Gough	F	47	13 cm	Tumor excision Sp, Adr	—	—
5	Banerjee and Gough	F	38	11 cm	Tumor excision Hy, Adn Colon	—	Mediastinal metastases 4 years after surgery
6	Park et al.	F	40	24 cm	Tumor excision Hy, Adn	Chemo	NER 3 months after surgery
7	Soendergaard and Kaspersen	F	37	18 cm	Tumor excision Hy, Adn	—	NER 18 months after surgery
8	Gotoh et al.	F	44	12.5 cm	Tumor excision	Chemo	Spread and death 4 months after surgery
9	Tenti et al.	F	46	20 cm	Tumor excision Hy, Adn	Chemo	NER 33 months after surgery
9	Tenti et al.	F	45	20 cm	Tumor excision Hy, Adn	—	NER 19 months after surgery
10	Motoyama et al.	F	42	11 cm	Tumor excision	—	—
10	Motoyama et al.	M	63	6 cm	Tumor excision	—	—
11	Carabias et al.	F	43	15 cm	Tumor excision Hy, Adn	—	NER 2 years after surgery
12	Lee et al.	F	55	19 cm	Tumor excision Hy, Adn	No	NER 30 months after surgery
12	Lee et al.	F	45	17 cm	Tumor excision Hy, Adn	No	NER 15 months after surgery
13	Pearl et al.	F	33	—	Tumor excision	—	NER 10 months after surgery
	Present case	F	86	23 cm	Tumor excision Gastrectomy	—	NER 6 years after surgery

*—, unknown; Hy, hysterectomy; Adn, adnexectomy; Sp, splenectomy; Adr, left adrenalectomy; Colon, colon resection; Chemo, chemotherapy; NER, no evidence of recurrence.

ectomy with bilateral salpingo-oophorectomy was performed in 11 cases, either simultaneously or later. Some reports [8,12] recommend total hysterectomy with bilateral salpingo-oophorectomy because recurrence was more frequent when simple tumor excision was performed; the tumor has been said to be influenced by female hormones. Eight patients have been reported to be alive without recurrence 3 months to 6 years after operation. Our case involves the longest known survival. Recurrences of the tumor developed in 4 cases [1,2,5,8] and 2 patients have died [1,8]. One case [2] recurred in the form of a left paraovarian cyst 21 months after a retroperitoneal tumor had been removed intact. The remaining patient [5] was alive with mediastinal metastases when reported. In a fatal case [1], in which the tumor capsule had been injured during operation, death occurred from distant metastasis. The other fatal case [8] involved death from disseminated tumor 4 months after resection. Adjuvant chemotherapy was administered in 4 cases [2,6,8,9], but its effectiveness is uncertain. Our case was the only one associated with mucinous peritoneal implants.

Dissemination or rupture of a mucinous cystadenocarcinoma at any site could lead eventually to pseudomyxoma peritonei. However, pseudomyxoma peritonei caused by retroperitoneal mucinous cystadenocarcinoma has not been reported. Our patient has survived for 6 years after operation without clinical signs of this complication. Similar to mucinous cystadenocarcinoma of other organs, retroperitoneal mucinous cystadenocarcinoma often grows slowly. Long survival therefore can be achieved following total removal, even with mucinous peritoneal implants.

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